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JUNE 2018 Thursday

d'Esch/Alzette Salle de Projection *

House of BioHealth, **Room Francoise** Barré-Sinoussi 11.30 am - 12.30 pm 1.00 - 2.30 pm

> *Please register sending a mail to florence.henry@lih.lu



SPEAKER Prof Klaus WARNATZ

Leader of Research Group "Human B-cell immunodeficiency"; Senior Physician, Medical Center - University of Freiburg, CCI – Center for Chronic Immunodeficiency, Freiburg, Germany

HOST: **Department of Infection** and Immunity

RESPONSIBLE LIH SCIENTIST:

Dr Jacques Zimmer (jacques.zimmer@lih.lu)

AUTOIMMUNITY IN IMMUNODEFICIENCY – WALKING ON A TIGHT ROPE

ABSTRACT

Autoimmunity is a leading clinical manifestation in patients with common variable immunodeficiency (CVID). About 30% of CVID patients suffer from overt autoimmunity and immune-mediated organ inflammation, which determine to a large extent the prognosis of these patients. Recent years allowed for better insight in the pathogenesis of this heterogeneous complication. While autoimmune cytopenias (Autoimmune hemolytic anemia and autoimmune thrombocytopenia) are predominantly B cell mediated, autoimmune enteropathy is mainly a problem of dysregulated T cell immunity. Interstitial lung disease seems to involve both arms of the adaptive immune system. Very little is known about the innate immune system in this context, but a strong

association with an increased interferon signature has been postulated by several groups. Several monogenic causes are associated with the clinical presentation of CVID with secondary non-infectious complications adding to the picture. Finally, the first manuscripts demonstrate the contribution of a dysbalance in the gastrointestinal microbiome to the immune dysregulation in the affected patients. Thus, autoimmunity is the other side of the coin of immunodeficiency in a substantial group of CVID patients. We are still seeking a full picture of this complex immune dysregulation affected by genetic and non-genetic components in order to offer better treatment or even prevent these complications.

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